




Spontaneous Rupture of Choledochal Cyst: A Rare Presentation

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Abstract

Choledochal cyst (CDC) is a rare surgical cause of cholestatic jaundice in infants. Spontaneous rupture is an unusual presentation of a previously undiagnosed CDC and is also a rare cause of biliary peritonitis in children. Here, we report a case of a 1-year-old boy who was evaluated for progressive abdominal distension. Ultrasonogram showed gross ascites with echogenic particles, dilated common bile duct (CBD), common hepatic duct (CHD), and upstream intra hepatic biliary radicle dilatation (IHBRD). CECT sections of the abdomen showed gross ascites and IHBRD with disproportionate dilatation of CHD and CBD. At laparotomy, a type 1 CDC with rupture of the anterior wall was found. The cyst was excised followed by hepaticojejunostomy. In a sick child with abdominal pain, cholestatic jaundice and biliary ascites, a high index of suspicion during imaging will help in the correct diagnosis and surgery for a potentially fatal ruptured CDC.

Keywords

- ▶ choledochal cyst
- ▶ rupture
- ▶ biliary peritonitis

Introduction

Choledochal cyst (CDC) is a rare surgical cause of cholestatic jaundice in infants. Spontaneous rupture is an unusual presentation of a previously undiagnosed CDC and is also a rare cause of biliary peritonitis in children. Here, we report a case of a 1-year-old boy who was evaluated for progressive abdominal distension. Ultrasonogram showed gross ascites with echogenic particles, dilated common bile duct (CBD), common hepatic duct (CHD), and upstream intra hepatic biliary radicle dilatation (IHBRD). CECT sections of the abdomen showed gross ascites and IHBRD with disproportionate dilatation of CHD and CBD. At laparotomy, a type 1 CDC with rupture of the anterior wall was found. The cyst was excised followed by hepaticojejunostomy. In a sick child with abdominal pain, cholestatic jaundice and biliary ascites, a high

index of suspicion during imaging will help in the correct diagnosis and surgery for a potentially fatal ruptured CDC.

Clinical History

A 1-year-old male child presented with vomiting, loose stools, and loss of appetite for 1 week. He developed progressive abdominal distention 2 weeks later. There was no history of fever, jaundice, or abdominal trauma. He had mild epigastric fullness but his abdomen was soft and nontender. His serum amylase was 550 IU (30–110), lipase 640 IU (10–140), alkaline phosphatase 980, total bilirubin 7, 5, and conjugated bilirubin 5.6. His Hb was 9 g%, total count 15,800, polymorphs 60%, ESR 88, and CRP110. A diagnostic ascitic tap was done that showed greenish bile-stained fluid, suggestive of biliary peritonitis (▶ Fig. 1).

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Fig. 1 Diagnostic ascitic tap showing greenish bile stained fluid (Dept. of pediatric surgery, Govt. Medical College, Kozhikode).

Imaging Findings

Ultrasonogram showed gross ascites (► **Fig. 2**) distended gall bladder, echogenic sludge in the distal CBD (► **Fig. 3**) with upstream dilatation of CBD, CHD (► **Fig. 4**), and IHBRD. CECT sections of the abdomen showed gross ascites and bilobar IHBRD (► **Fig. 5**) with disproportionate dilatation of CHD and CBD (► **Fig. 6**). Gallbladder was distended with normal wall. There was biliary sludge in distal CBD and an ill-defined walled off collection in the left subhepatic location, which was communicating with the anterior wall of CBD (► **Fig. 7**), suggestive of choledochal cyst rupture with biliary ascites.



Fig. 2 Ultrasonogram of the abdomen showing gross ascites with echogenic particles (red arrow).

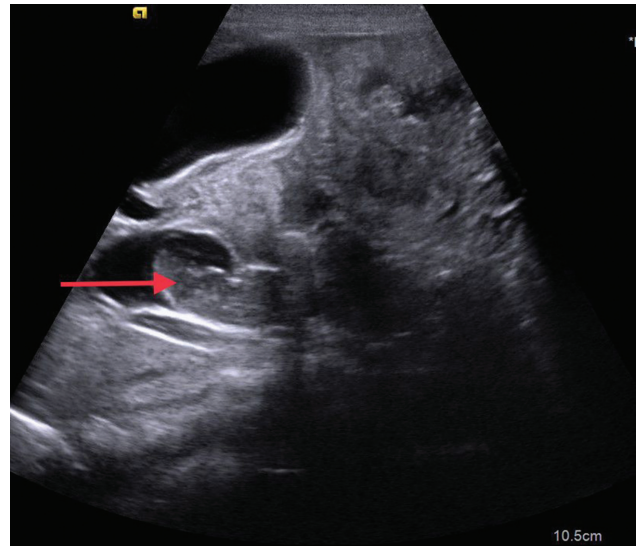


Fig. 3 Ultrasonogram of the abdomen showing echogenic sludge in dilated common bile duct (red arrow).

An exploratory laparotomy was done that revealed a type I CDC with perforation of the anterior wall (► **Fig. 8**). Excision of CDC and hepaticojejunostomy were done.

Discussion

Choledochal cysts are congenital bile duct anomalies in which cystic dilatation of the biliary tree occurs in the extrahepatic biliary radicles, intrahepatic biliary radicles, or both. It is due to congenital weakness of the duct wall, or defective development or weakening of duct walls due to reflux of pancreatic enzymes in case of anomalous development. CDCs are rare (1/100000 and 1/2000000 live births) and are more common in females (female-to-male ratio: 3:1 to 4:1). They are more common in Asian countries especially Japan.^{1,2} The classic triad of CDC, intermittent abdominal

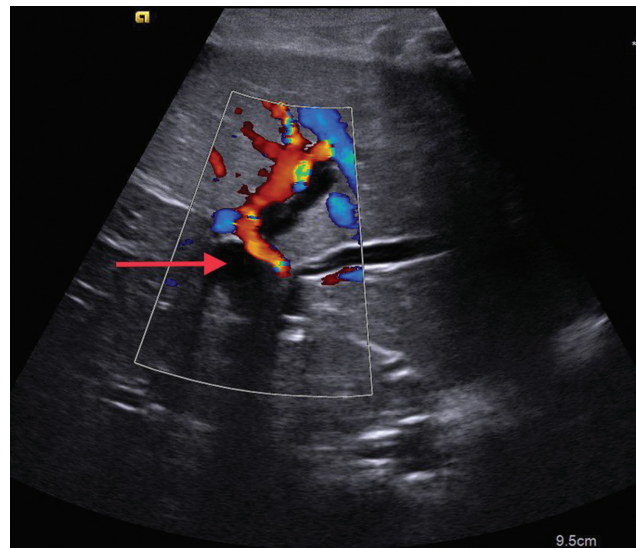


Fig. 4 Ultrasonogram of the abdomen showing dilated common hepatic duct (red arrow).

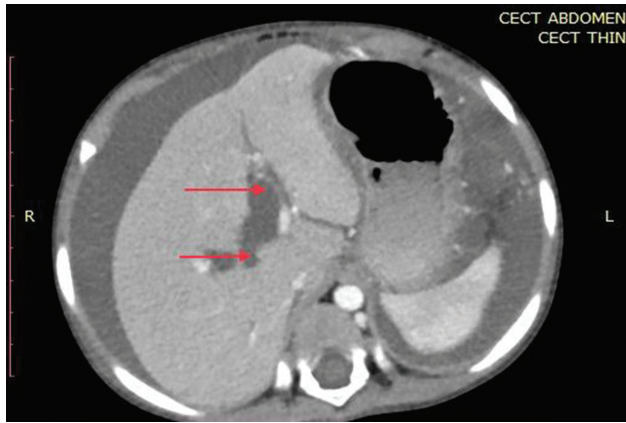


Fig. 5 Axial CECT abdomen showing bilobar intrahepatic biliary radicle dilatation (red arrows).

pain, jaundice, and a palpable right upper quadrant mass, is more common in children than adults. However, most patients will have only one or two elements of the triad.³⁻⁵

Todani classified CDC into six types in 1983. Type I cyst (50–85%) is characterized by cystic or fusiform dilatation of the CBD with normal intra hepatic ducts. Type II cyst (2%) is an isolated CBD diverticulum. Type III cyst or choledochocele (1–5%) is a cystic dilatation limited to the intraduodenal portion of the distal CBD. Type IV cysts (15–35%) are divided into IVa with multiple dilatation of the intrahepatic and extrahepatic biliary tree, and IVb with multiple dilatation involving only the extrahepatic biliary tree. Type V cyst or Caroli disease (20%) is characterized by one or more cystic dilatation of the intrahepatic ducts, without extrahepatic duct disease. Type VI cyst, isolated cystic dilatation of the cystic duct, is very rare.⁶⁻⁸ Children with CDC may present in the neonatal period with cholestatic jaundice that may be mistaken for biliary atresia. If not diagnosed and treated promptly, these children may develop recurrent cholangitis, pancreatitis, cirrhosis, portal hypertension, variceal bleed and ascites.⁹ Unoperated children with CDC, especially types 1 and 1V, are prone to develop cholangiocarcinoma later.¹⁰ A

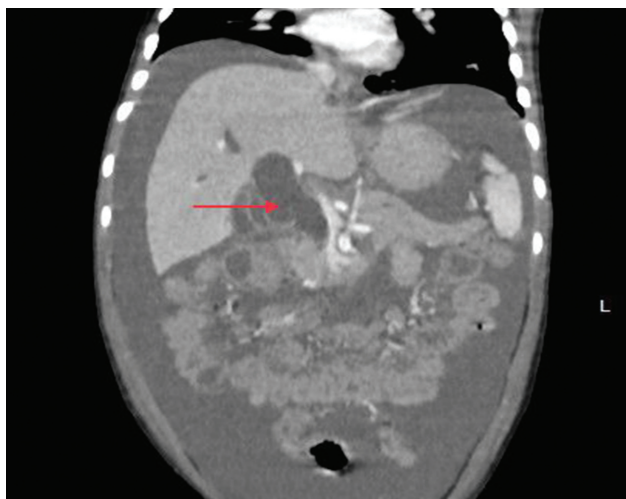


Fig. 6 Coronal reformatted CECT abdomen showing fusiform dilatation of common bile duct (red arrow).

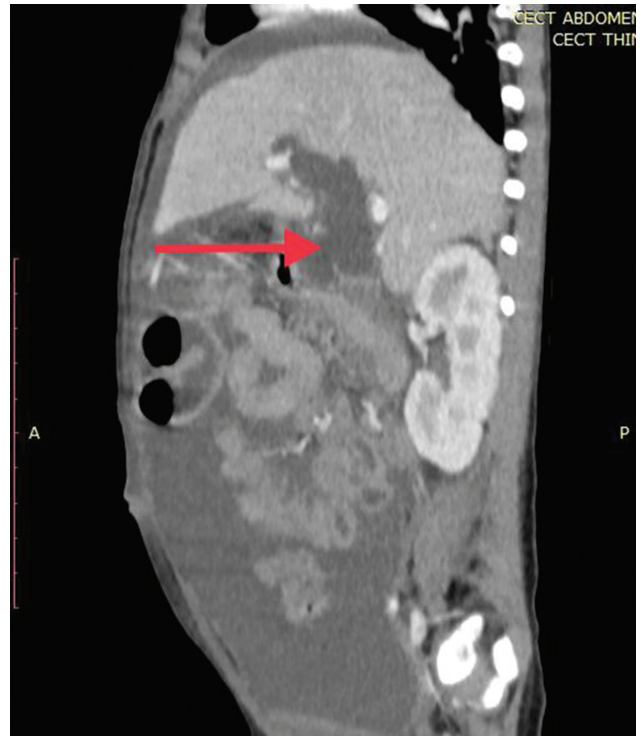


Fig. 7 Sagittal reformatted CECT abdomen showing defect in the anterior wall of dilated common bile duct (red arrow).

rare presentation is as an acute abdomen due to rupture in 2–18%.¹¹ Complete resection is the optimal therapy.¹²⁻¹⁴

This child with an undiagnosed CDC had a dramatic presentation as spontaneous rupture. Rupture of CDC is

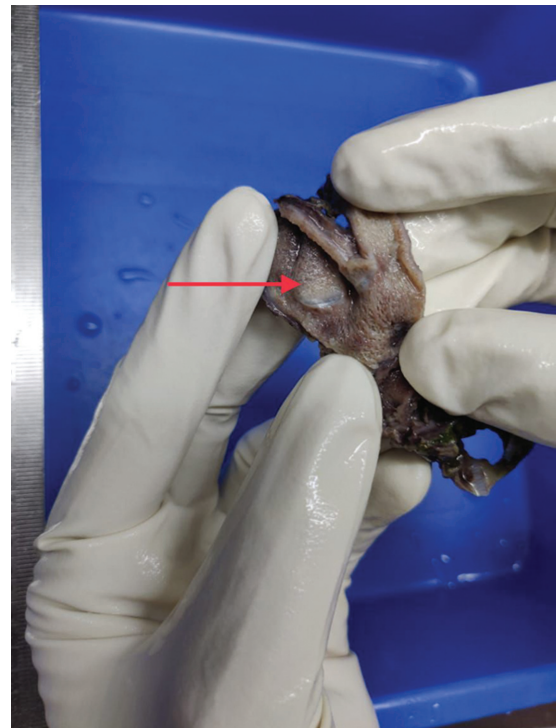


Fig. 8 Gross pathological specimen showing cut open dilated common bile duct (red arrow) (Dept. of Pathology, Govt. Medical College, Kozhikode).

usually seen in neonates following some precipitating events such as trauma, which were not seen in our patient. Spontaneous rupture presents as biliary peritonitis with features of sepsis.¹⁵ Some of the postulated mechanisms of rupture include abdominal trauma, pregnancy, labor, distal obstruction, epithelial irritation by reflux of pancreatic secretions, and wall necrosis. However, the exact etiology remains obscure in many cases.^{11,16-18}

Choledochal cysts can be diagnosed by ultrasound scan, which shows disproportionate dilatation or cystic lesion of bile duct. CT/MRI can be used to assess the intrahepatic disease and complications.¹⁹ The differential diagnosis includes dilated CBD due to stricture or calculi. The mainstay of treatment is surgical resection in both ruptured and unruptured cysts. If total resection is technically difficult, partial resection is the procedure of choice.^{20,21} Spontaneous rupture of CDC should be suspected in all sick children with cholestatic jaundice, abdominal pain, and biliary ascites.

Unfortunately, this child had a stormy postoperative course, developed florid septicemia and expired on the third day despite appropriate antimicrobials and supportive measures.

Conclusion

Bile duct cysts are congenital dilatations of the intra-hepatic and/or extra-hepatic biliary tree. Todani divided these cysts in five groups. Known complications of bile duct cysts include lithiasis, cholangitis, and malignancy. Spontaneous perforation with biliary peritonitis is a rare complication of bile duct cysts, described in less than 2% of cases, usually in children. The diagnosis can be suspected in a patient with acute abdomen, ascites, and dilated biliary tree on imaging. CT or MRCP can show the perforation site in the cyst wall. Timely diagnosis if necessary for proper management and is life-saving in these cases.

Conflict of Interest

None declared.

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